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ABSTRACT

The study examined aspects of daily living of Swedish children with osteogenesis imperfecta, a mineral deficiency in the skeleton which results in stunted growth and frequent fractures. A questionnaire was administered to 24 families with children under the age of 18 and 3 families were interviewed. The study found the families in great need of support services. Early diagnosis is particularly important to prevent families from being accused of child abuse. Professionals and authorities were found to lack sufficient knowledge about the condition. A need was found for modifications of the children's homes and immediate environments and for suitable handicap aids. Families were found to need both psychosocial support and practical help with modification of the home and obtaining of aids. The report includes a discussion of the daily life and modifications made in the three families interviewed. Four references (not in English). (DB)

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CHILDREN WITH OSTEOGENESIS IMPERFECTA AND THEIR DAILY LIVING

JANE BRODIN

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Handicap Research Group
Report No. 4, 1990

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JANE BRODIN

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3

UNIVERSITY COLLEGE OF JÖNKÖPING
HANDICAP RESEARCH GROUP

ABSTRACT

JANE BRODIN
THE WRP GROUP

CHILDREN WITH OSTEOGENESIS IMPERFECTA AND THEIR DAILY LIVING

Report No 4, 1990 ISSN: 1100-9047

The aim of the study has been to describe the daily living and the state of arts in the area of handicap aids for children with osteogenesis imperfecta. The study is part of a larger research project on the situation for families of children with osteogenesis imperfecta, initiated by RBU (the National Association for Disabled Children and Young People).

The study is based on a questionnaire administered to 24 families with children under the age of 18, and interviews with three families. The study shows that the families with children suffering from osteogenesis imperfecta are in great need of support. It is important with an early diagnosis to prevent the families from being suspected of child abuse. Professionals and authorities often seem to lack sufficient knowledge about the condition and its implications. Another important area is modifications of the home and the immediate environment. In addition there is a great need for knowledge and information in the area of handicap aids. Professionals at the handicap aids centres most often know little about the condition and suitable aids for those children. Thus the families need both psychosocial support and concrete help with modification of the house and environment and handicap aids.

Key words: Osteogenesis imperfecta, brittle bones, children, psychosocial support, handicapaids, home modifications.

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FOREWORD

Children with osteogenesis imperfecta (OI) form a small and relatively unknown group of handicapped people. These children and their families are often faced with complex problems and they need a great deal of help and support. Because this condition is so rare, knowledge and experience of osteogenesis imperfecta is very limited. It is estimated that less than 300 people in Sweden suffer from this disease.

I should like to express my warm thanks to Maria, Teres and Tommy and their families, who welcomed me into their homes and allowed me to learn from their knowledge and experience. I should also like to thank Viola Larsson and Gunilla Bragsjö of the OI working group, the National Association for Disabled Children and Young People, for their cooperation and assistance.

I hope that this report will be of benefit to parents and others who deal with children with osteogenesis imperfecta and their need for special assistance and support.

May 1990

Jane Brodin

Handicap Research Group
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1 INTRODUCTION

1.1 Background

About five children born in Sweden each year are diagnosed as suffering from osteogenesis imperfecta (OI). A symtom of this condition is that fractures are easily sustained in every part of the skeleton. Children with osteogenesis imperfecta also have weaker than normal tendons, sinews, muscle attachements and other elastic tissue. A blue tint in the whites of the eyes and stunted growth are also common symtoms. However, these children do not suffer from any form of brain damage.

There are probably around 100 people in Sweden under the age of 17 suffering from osteogenesis imperfecta. It is impossible to give an exact figure because no detailed survey has been made in Sweden since the 1950s, when Dr Gunnar Smårs carried out a survey which was, however, not fully comprehensive. It is common that OI sufferers sustain numerous fractures, and their limbs are often so deformed that they become confined to a wheelchair. There is a considerable need for information and technical aids, and for the provision of specially modified appliances and apparatus to enable these children to develop satisfactorily.

1.2 Objectives for the study

This project aims to make an inventory of the need of technical aids and modifications for children with osteogenesis imperfecta. Another aim is to describe the families' life situation. The target group for this work consists of parents, occupational therapists, physiotherapists in child habilitation, midwives and child care staff.

1.3 Method

A questionnaire was sent to all families registrated with the Society for osteogenesis imperfecta, a branch of the National Association for Disabled Children and Young People (RBU). A total of 38 families answered the questionnaire, and three of these families were also interviewed. A reference group, which included Gunilla Bragsjö and Viola Larsson from the RBU, were involved in the project. The results of the questionnaire and the interviews are presented in this report.

2. WHAT IS OSTEOGENESIS IMPERFECTA?

The latin name osteogenesis imperfecta means imperfect or incomplete skeletal formation. The disease is a very old one; history describes people of glass china, indicating that they were very brittle. A mineral deficiency in the skeleton is common to the two types of osteogenesis imperfecta. A defect has also been found in the binding tissue of a protein called collagen. In recent years, amino acids deficiency has also been found to be a decisive factor in the occurrence of this condition. Osteogenesis imperfecta is an inherited disease, but the condition can also appear in a family which has never had this disease. This may be caused by a latent hereditary disposition for the disease, or by a genetic mutation.

The national association for motor disabled children and young people has the following view of the symptoms of the two main types of osteogenesis imperfecta; congenita and tarda.

Osteogenesis imperfecta congenita - patients are born with unmistakable symptoms. Congenita is the rarest type of the disease and is usually inherited. Some children are so seriously affected by the disease that they die in utero or very short after birth. Many of the surviving children are born with unhealed or badly healed fractures.

Osteogenesis imperfecta tarda - the symptoms occur at a later stage, as a rule after the age of 18 months - two years. There is a severe form of tarda, with many fractures and serious deformations, and a less dominant inherited form with fewer fractures, and which does not usually cripple the sufferer.

The following summary of the symptoms focuses primarily on osteogenesis imperfecta congenita and the more serious forms of osteogenesis imperfecta tarda.

1 The fractures, which are just as painful as in children with normal skeletal formation, occur most commonly in the arms and legs, but the back, ribs, collar bone and fingers can also fracture. The fracture may be caused by a mere muscle movement. Most of the children have very stunted growth; the lower legs and thighs are bent; the sinews, tendons and other elastic tissue are weak and the muscles are thin.

2.There is often blue coloration of sclera, the hard covering of the eye, because it is thinner than normal.

3.The teeth often develop late. In a few children, the teeth are soft, the enamel cracked, and the teeth worn down to the gum (dentinogenesis imperfecta). Brittle teeth which very easily break off are a more common symptom.

4.Later in life, as a rule not before the age of 25-30, about half of the children middle ear. This increasing deafness may be reduced or corrected by surgery or the use of hearing aids

5.The blood vessels are often fragile and the children bruise easily.

6.At birth, the cranium is soft. Many children have a triangular shaped face with a broad forehead and narrow chin. However, this condition does not affect the brain or the brain cells. Children with osteogenesis imperfecta enjoy normal mental development.

7.Many children have a slightly higher body temperature, perspire more than normal and need a higher than normal fluid intake.

3. THE QUESTIONNAIRE SURVEY

A total of 38 questionnaires were sent out to everyone registrated as members of the RBU group for people with osteogenesis imperfecta. Two questionnaires, each with a different set of questions, were sent out, one to parents of children under the age of 18 (a total of 27) and the other to adults with osteogenesis imperfecta (a total of 11).The addresses were obtained through the RBU work group for osteogenesis imperfecta, and the group has also submitted aspects on the questions included in the questionnaires. Thirty-two responses were recieved. Only 6 families failed to complete and return the questionnaire.

Only the findings concerning children under 18 are reported in this study (24 families). The children of those families, 15 boys and 9 girls, were born between 1968 and 1984. Six of them were the only child in the family and 18 were from families with two or three children. None of the brothers and sisters have osteogenesis imperfecta, but one parent suffers from this condition.

3.1 Informing the parents

Thirteen of the parents were told when their children were born, that they had osteogenesis imperfecta. All the other parents were told before the children had reached the age of three, as set out in the table below:

At birth	13 parents.
0 - 6 months	2 parents
6 - 12 months	5 parents
1 - 3 years	4 parents

All parents were given this information verbally, and in 20 cases they were told by a doctor. More than half of the parents were not given a good picture of the implications of the handicap. Only three parents say they were given a comprehensive picture of the difficulties involved. Four families were helped at an early stage to

establish contact with another family who also had a child suffering from osteogenesis imperfecta. The parents say that the reason why so few of them were given help was that the professionals did not know enough about the condition and its implications.

3.2 Daily activities and play

Seven of the children are integrated into the preschool/day nursery system, five are at school and the rest are at home with their parents. 14 of the children have playmates who come home and play with them, while the others have noone to play with. The parents are often worried that the children break their bones when playing with their friends. More than half the parents (13) say that they are often worried, and the others say that they are worried at times. In response to the question of whether the children themselves are worried about fractures, only four parents answered yes, while the others answered sometimes. Three of them said never. One parent answered "don´t know" and one gave no answer because the developmental age of the child made the question irrelevant.

3.3 Hydroptheraphy

Hydrotherapy is a very good way of building up the physical strength of osteogenesis imperfecta sufferers, and they place considerable value on this training. Nineteen families have access to a pool, and in nine cases the pool is a municipal bath. Three families have a pool at home and seven families use a pool either at school or at friends´ homes. Only some of the families think that hydrotherapy works satisfactorily or well. Five families find this a problem and the parents feel that a home modification grant to build a pool at home would be the best solution.

3.4 Fractures

Children with osteogenesis imperfecta often suffer broken bones. In several cases authorities raised the question of whether these injuries were caused by child abuse. The parents came under this kind of suspicion in eleven of the families, while the same number of families have never been suspected of child abuse. In two cases, the social welfare authorities were alerted. In one of these cases the child was placed in a foster home for a month. In the other case the authorities realized what the situation was in time, so the child was not mistakenly taken into care.

One little girl was 16 months old when osteogenesis imperfecta was diagnosed. The parents felt that they were under constant suspicion during the three months it took to complete the investigation of the case. One boy was diagnosed at the age of 10 months. Prior to this he has been the subject of a seven-month long investigation, throughout which the parents felt they were suspected of child abuse.

In the past year the children have suffered between one and five fractures each on average. Having lokeed at previous records of fractures among these children we chose a scale between 10 and 100 breakages. Only one child (born in 1983) had suffered as few as three fractures.

3.5 Technical aids

Ten of the children use standard equipment to help them, and five have individually-modified appliances. A combination of standard and modified equipment seems to give the best results. The following answers were given to the question of whether the families had been given the handicap aids they required:

12 families answered yes

5 families answered no

6 families answered don´t know

1 family gave no response because they had no appliances (child born 1983)

Eleven of the children´s aids were functional and worked well and nine worked fairly well, i.e, they were not entirely satisfactory but good enough under the circumstances. The other children´s aids were not satisfactory.

In response to the question of whether there was a need for equipment which had not yet been supplied, ten families answered that there was such a need, while the same number answered that all their problems had been solved. The parents stated that at present there were no handicap aids available in the following areas: mobility, lifts, communication, soft, shock-absorbing wheelchairs (both manual and powered by electric motors), walking frames, personal hygiene aids, car seats, apparatus for exercising while lying on the stomach, various kinds of training apparatus, (for keep,fit exercises etc.), leisure time aids for country walks and skiing, housing modifications and modifications to the environment (e.g. the entrance and approach to the home).

It should be possible to develop most of this kind of equipment for general use and then make individual modifications. This is a difficult area for the staff at the handicap aids centres because they are not very familiar with the problems associated with osteogenesis imperfecta.
According to the responses, equipment is needed in the following areas:

Personal hygien	5
Lifting equipment	5
Sitting aids	7
Transportation	11
Communication	10

No matter where the osteogenesis imperfecta sufferer goes, the problems are considerable. Half the families say that the problems are just as big at home as they are at school and in leisure-time activities. Some parents feel that the most outstanding problems are related to leisure time activities because the child cannot take an active part in "social life".

3.6 The parents' view on handicap aids

At the end of the questionnaire the parents were asked to give their own views of their problems when it comes to equipment. The following quotations are from the parents´ comments:

- Delivery times are too long.

- We need financial assistance for various kinds of technical aids and the authorities should be more responsive to our needs.

- We need a smoothly-operating system with shorter waiting periods for all kinds of equipment.

- We would like to have easy-to-use practical aids.

- I don´t think that as a parent one should have to work out what aids the child needs. I have never been able to borrow a brochure on available appliances and equipment from the handicap aids centre. You should be given one automatically when you need this equipment.

- The "Frontal" bed for children to exercise on their stomachs would be perfect if it could be made a size smaller.

- There are some problems associated with exercises to build up strength, particulary for the legs. An exercise cycle is available in a children´s model might solve our problems.

- The bath could be replaced with a small, deeper pool with straighter sides. This could be used for practicing walking. It should be fitted with a door so that the children do not have to be lifted in and out.

- As children are so different, I belive that the suggestion which are suitable for some children may need individually modifications to suit others.

- The occuptional therapists do not know enough about what is available on the market. They are also not very familiar with osteogenesis imperfecta and this means that the parents have to find out for themselves what aids are available and then suggest them.

- The need for aids varies considerable from case to case. What is needed most is to have information about the circumstances of osteogenesis imperfecta sufferers circulated to children´s clinics, the handicap aids centres, the preschools, schools and various agencies.

4 INTERVIEWS WITH THREE CHILDREN AND THEIR PARENTS

Interviews were carried out with three children and their parents with the aim of getting a picture of the daily living conditions of children with osteogenesis imperfecta, and in what way their handicap affects the life situation and circumstances of the family. The three children are Maria, aged 13, Theres, aged 8, and Tommy, aged 5. These children come from different parts of Sweden and, like most disabled children, attend the ordinary municipal day-care facilities and go to an ordinary school.

4.1 A meeting with Maria

Maria is 13 years old. She lives with her parents and a three year older brother in a detached house in a small town in southern Sweden. When Maria was born her parents were told by the doctor that she had osteogenesis imperfecta, i.e., that her bones were brittle. The parents were given a fairly good description of the implications of this handicap, but the staff did not realize that it would have been very helpful if the parents had been put in touch with another family which had experience of osteogenesis imperfecta or which had a sufferer in the family. This would have helped to lay many worries to rest and answered many of their questions.

Maria goes to an ordinary school in which also two other children with disabilities attend, although they are not all in the same class. The school has been properly modified to suit disabled people; it has single-storey buildings and many open, easily-accessible areas. Being integrated into the ordinary school means that Maria also has friends outside the school. Her mother is often worried that her bones will break when she is playing with other children. But Maria herself is only occasionally worried that her friends might forget themselves at times and bump into her a little too hard and fracture her bones.

As the family was told about Maria´s osteogenesis imperfecta at birth, they have never felt that they were being accused of child abuse, as was the case with many other parents. However, Maria has many difficulties. The most important thing for her at the moment is to find clothes that are suitable yet fashionable. Clothes are often a passionate interest for girls in their early teens.

Few modifications has been made to make Maria's immediate neighbourhood suitable for disabled people. This causes Maria problems, partly because she is short, and partly because she is hurt by bumps and shakes.

When it comes to handicap aids, Maria almost always uses standard apparatus which has been specially-modified, and for that reason the aids she uses today work fairly well. But there are needs for equipment which are as yet unsatisfied. One major problem is that there are no manual or electrically-powered soft, shock-absorbing

wheelchairs. Moving from the wheelchair to the bed, the toilet or to another wheelchair is another problem.

Many of the problems people with osteogenesis imperfecta face require equipment which is in fairly wide use but which needs to be specially modified. The handicap aids centres usually know nothing or very little about osteogenesis imperfecta and the staff merely do the work the parents ask for. Maria´s parents say that there is no equipment in the fields of personal hygiene, mobility and transportation, for communications or for lifting Maria. The need for equipment and appliances is equally acute in the home, at school and for leisure activities. The family has a minibus which has been modified to suit Maria´s handicap. This was financed by the local and county authorities. Maria sits facing the back of the bus in a specially-modified "driver´s seat". The seat is electrically heated and has been fitted with a reading light. An extra floor has also been fitted to the luggage space, with clamps for a Permobil wheelchair. The passenger seats are fitted with specially-made protectors to stop unwanted movement and there are two telescopic ramps located in the space between the floors which can be pulled out to load and unload her wheelchair.

Turning to equipment and appliances, the family feel that the occupational therapists know very little about what is available on the market. This, combined with an insufficient understanding of osteogenesis imperfecta, means that the parents themselves have to find out what is available and give suggestion to the professional responsible for prescribing equipment and apparatus. Unfortunately, the occupational therapists do not have enough time to follow up prescribed appliances to see if they are well suited to the user. The long delivery times at the handicap aids centres is another problem.

Maris has suffered 25 fractures, which is an average of two a year. She has had no fractures for the past year, which indicates that she now knows a lot about her condition and is beginning to be able to deal with it. In other words, she has become aware of her handicap. About a year ago she had surgery to insert metal pins into her femurs to stop them breaking. This has been a great help.

4.1.1 Home modifications

When Maria was six years old, her parents built the detached home where they now live. It is a well-planned bungalow which has been modified to the requirements of a teenager and adults. Because they knew that Maria would be below-average height they installed wide doorways and bumpers to stop doors closing unexpectedly, and doorways without floor lintels. They also fitted double light switches (one low and one at normal height), a separate toilet and a specially-modified room to meet Maria's personal hygiene requirements. In this room Maria has a low-level hand basin with a shelf and mirror and a toilet fitted with a Hea toilet inlay seat. There is also shock-absorbent floor covering, specially-laid approach paths outside the house and a separate 11 square metre store for her equipment. The county housing committee gave a grant for these modifications.

Hydrotherapy in a swimming pool is an important part of the rehabilitation of people with osteogenesis imperfecta because this training strengthens the muscles and reduces the occurrence of fractures. Nowaday it is generally thought that fractures are less painful for people suffering from osteogenesis imperfecta than for other people. But fractures are always painful. Knowing the importance of hydrotherapy in strengthening muscles, the family have applied for a grant to install a 3 x 5 m indoor

pool. However, the housing board turned down their application with the motivation that the installation "was related to rehabilitation and functional training", and therefore not eligible for a grant. However, the family financed a pool themselves, and an annual grant is paid to heat the pool to a temperature to 32 C.

A major problem is that Maria cannot move from her chair to the floor, from her bed to her wheelchair and from her wheelchair to the toilet. Her arms are strong, so she should be able to move unaided because but these moves must be risk-free, because if she falls she may fracture a bone. However, she carries out her personal hygiene routines unaided.

4.1.2 A day with Maria

When Maria wakes up in the morning, she must receive help to get out of bed and move from her bed to the wheelchair in which she goes into her special bathroom. After washing and dressing herself, Maria has breakfast in the family kitchen. Maria sits at the kitchen table in a work chair for children, which is fitted with a special board. The chair is height-adjustable and fitted with an extra foot support.

Maria cannot make her own breakfast because she cannot reach the fridge or the hob. When she has finished her breakfas her mother moves her to her wheelchair so that she can go and brush her teeth. Then her mother lifts Maria down and puts her on the floor, and Maria puts on her boots and outdoor clothes. This is hard work for Maria and takes a long time, but she does it herself because she wants to be as independent of other people as possible. Maria has a personal assistant who has been with her since play school. When she is ready for school the assistant takes her to school. Maria switches wheelchairs and drives her cerise-coloured Permobil. Maria is ´individually integrated´ into a class of 19 pupils. She takes part in all the school activities and her school friends both accept and respect her. She is often left behind when the other children rush off to do something, and she would like to be able to keep up and be like the other children. The older she gets, the more difficult this becomes.

Sometimes Maria plays with the girl next door who is a year younger than her. "Her classmates often go down to the town after school to ´meet boys´, and she realizes she can´t go with them", says Maria´s mother. Maria has a contact person - a 20-year old girl - who she meets once a week for leisure activities. They usually go to the cinema or a disco together. She has had the same contact person for two years now and they get on very well together. They do not meet on any particular day, burt agree on a day which will suit them both. Because Maria reads a lot they also visit the local library together to borrow books.

When Maria comes home from school she parks her Permobil in the garage and her mother carries her indoors and puts her on the floor in the hall, where she takes her outdoor clothes off.

4.1.3 Maria's interests

Maria is interested in singing and music. She sings in the school choir and plays piano at the municipal music school. She has a varied taste in music, but at the moment she is most interested in rock music. Her favourites are "Europe" and "A-ha". She is also very interested in reading and gets through a lot of books. She reads 1-2 books a day, usually romantic stories. She tells us that she has been to the cinema

and seen "Karate Kid II". This is a love story which she liked very much. She thinks it is the best film she has ever seen. Just like most other young people, Maria likes to go to discotheques and dances. She is very interested in smart clothes, jewelry and make up.

Maria has spent a holiday abroad with her parents. Going abroad with Maria was a bold step for her parents. "You must tell all young people that you can travel abroad even if you are in a wheelchair", says Maria. You can see from her expression that the holiday was a wonderful and valuable experience for her. All the potential problems which had given her parents and, to some extent even Maria, sleepless nights before the holiday, had been solved. The family were given good service and everything went very well.

When Maria grows up she would like to be a librarian or perhaps take an active part in the work of the RBU, of which she is a youth member today.

4.1.4 Maria's handicap aids

Maria has a table fitted with wheels. It has a crank to adjust the heigth. Next to the table there is a **Brio Rehab** wheelchair. It has been specially modified, with a raised foot support and a shorter seat. Maria often sits in it to do her homework.

In the evenings it is pleasant to sit in a comfortable foam rubber armchair. Maria bought it at Ikea, the furniture store. The armchair has no legs but stands flat on the floor so that Maria can climb in and out of it without any risk.

The 70 cm long **Hea Rehab wheeled board** is another aid which works very well. When Maria was smaller this was almost the only apparatus she used, particularly outdoors in the summer. It is very easy to manoeuvre.

It is important for Maria to move around by herself. She has a walking frame with a specially-modified seat sling. The gusset is much narrower than in the standard model, which gave her blisters. She uses this frame to make it easier to get close to wash basins and tables. In fact, Maria sits back-to-front.

Maria has also a **Spinner Kit wheelchair** and a cerise and pink **Permobil**. When she rides in it she looks proud and confident. Maria´s mother says that they have never had any problem about repainting the wheelchair, but she knows families who have not been given permission by the handicap aids centre to make any changes to the basic yellow model Permobil. This is a very important factor for Maria, and presumably for other children too, because they want a colour they like. A mobile telephone has been ordered from the centre and this will be a good safety precaution for Maria. Sometimes the Permobil breaks down and cannot be started. Because Maria cannot move unaided, and only in an emergency should she be lifted by strangers, this is a major problem. Maria will feel better provided the extra security of a mobile telephone.

Maria cannot make her own breakfast because she cannot reach up to the fridge, but she can open the fridge door. Maria says that she has now found a way of opening the fridge door herself. One day there was a large box of chocolates in the fridge and, necessity being the mother of invention, when Maria was desperate for a chocolate

one day she fetched her tongs and with a lot of effort managed to get the fridge open and lift down the chocolate. Happy she was!

One problem which has been solved is moving from one chair to another, i.e. Maria is able to use a lift herself. Solving this problem has given her greater oppurtunities to become independent and feel free and unrestricted. By using the lift, Maria can move around and no longer has to ask her parents for help. This is a considerable relief both for Maria and her parents.

4.2 A meeting with Teres

Teres, who is now eight years old, came to the her family as a foster child when she was eight months old. She could crawl then, but was small and thin. She often whimpered and was fretful. Her parents thought that she was developing slowly and felt that the fact that she was so small and thin explained why she had so little energy. Her parents began to train her to help her achieve a normal rate of development. She made considerable progress and gradually learned to both sit and walk unaided.

At 12 months, she stood up to walk but slipped and fell and hurt her neck. At fist her parents thought that she had a sprain, but when she did not respond to treatment with different ointments they took her to hospital. She was found to have fractured a neck vertebra, but there was nothing else wrong with her.

Some time later, another foster child joined the family, a boy one year younger than Teres. A month after he arrived, Teres fell over and hurt her arm. When a medical examination revealed a broken arm, the suspicion of child abuse arose. Mother and daughter went into hospital for an examination during the midsummer holiday but the doctor said nothing about suspicions of child abuse. No inquiry was carried out and after a week Teres contracted mumps. However, the authorities "checked" the family everyday to make sure that nothing should happened.

After living at home for two weeks Teres broke a rib, and the authorities decided that Teres and her brother should be brought in for a full x-ray examination. The doctors provoked the parents to anger them and catch them off guard. The social welfare officers thought the parents might have mistreated the children without realizing it. These things happened in their experience. The parents were reported to the police suspected of child abuse. However, two days before the scheduled interview with the police, Teres broke her leg at the hospital when another child picked her up and dropped her on the floor. If this incident had not occurred, the staff would probably still have suspected the parents of mistreating their children since there were a.medical statement of child abuse. While Teres' brother was in hospital he contracted a stomach infection. The effects of this illness lasted a long time, and and had to stay in hospital. This probably prevented the children from being taken from their parents and put into a children´s home.

Today, the parents try to encourage contact with their foster children´s biological parents. Tobias is adopted, and Teres is a foster child. Teres´ mother was very young when she gave birth to Teres and she was quite unable to look after her. Teres meets her mother now, but has no contact at all with her biological father.

4.2.1 Home modifications

When Teres came to the family they lived in a small 2-storey house with a staircase. Later on the family built a completely new single-storey house about 15 km from their first home. They did not know that they could apply for grants to modify their home, so they tried to manage as well as they could by themselves. They sent for a lot of brochures and designed the house themselves. Nobody from the local authority asked them if they needed any help with this job. They used to live in a different municipality, and the occupational therapist there helped them to apply for a home modification grant. They didn´t get very much; there was no ramp at the driveway entrance, and the paths outside the house had not been hardsurfaced. But they did get an extra shower room.

The family applied for a grant to build a 3 x 6 metre pool in their house. The application was rejected, first by the municipality, then by the county housing board and the national housing board, and finally the matter was put before the Supreme Administrative Court. A petition was sent to the Ministry of Housing in April 1986, and in the spring of 1987 they approved the application. The municipality was prepared to grant a Skr 20,000 home modification grant, but any work in excess of this amount was counted as an investment to improve the standard of the house and would therefore not be covered by a grant. The Supreme Administrative Court heard this matter in the summer of 1987, and ruled in the family´s favour. They got their pool.

The main modifications to the home have been to produce a bungalow with large open areas. There is a great deal missing, including a store for handicap aids - all the apparatus must be kept in the hall and the family normally uses the garage entrance. However, this is much better than their previous arrangements, and they are quite satisfied.

4.2.2 A day with Teres

Teres wakes up and gets out of bed at 6.30 every day. Because she has just been given a water bed she can no longer change and have her morning wash in bed. Teres´ mother changes her on a table in the room. As little as possible is done because it is painful for Teres, and many of her clothes are chosen with this in mind. If she has a lot of pain she chooses clothes which are easy to put on. As a rule, dressing Teres is hard work, taking about half an hour. Sometimes she choses her clothes herself, but her mother often helps her. Her favourite colour is pink, as can be seen from the decorations in her room.

The family has breakfast between 7 and 7.30. Teres´ arms are too weak for her to eat without help, so her mother has to hold her mug of gruel. Then Teres´ home help assistant arrives and they go to school. She has now had the same assistant for two years and the system works well.

The school has been modified to meet Teres´ needs. It is an old school, the same one that Teres´ father attended as a child. A ramp has been built so Teres can get into the school in a wheelchair. A hoist has been installed to Teres to move from the ground floor to the dining room on the first floor. Every day Teres and a friend leave for the dining room a little early. For safety reasons, she is allowed to leave before the other pupils, because she has to be lifted.

Sometimes her friends refuse to let Teres join in the play. They may tell her that they have decided to play by themselves and she is not allowed to participate. The other children move rapidly from place to place and Teres often cannot keep up. Just now Teres´ best friend is a girl in the same class.

Teres is taken home in the afternoon by the municipal school transport service. Once home, she usually rests on a mattress on the floor and lies there stretching her body for an hour or so. She strenghtens her arms with the help of a clothes hanger. Sometimes she watches a video when she is lying on the mattress and sometimes she eats a sandwich and drinks a little milk while she is resting. By this time her playmate has usually arrived and is sitting and waiting for her. Sometimes she telephones a friend who comes over to play. There are not very many children in the area so she usually calls one particular girl. It is easier to meet her friend now because she has got a bicycle of her own.

4.2.3 Teres' interests

Teres is interested in dance and music, clothes and make up, "and boys of course", she adds. She does not read very much because she finds it hard work. She likes music very much and Herreys and Style are her current favourites. She would dearly love to play the drums or electric guitar.

Teres has many and varied interests, she sings in the church choir one day a week, goes to a philatelists club every second week and to the brownies once a week. She also enjoys drawing and going to the cinema.Outdoors, her favourite pastime is to play in the hut and the play room that her father has built her, or with her little rabbit, Stampe. Indoors, she enjoy playing with small toys. Teres´ friend likes to play hospital, and rarely wants to go out, also preferring to watch a video or play arcade games on the TV. She does not like to play with dolls. "She always wants to order me about", says Teres, "and that can be difficult at times".

Teres wakes up screaming every night. When her mother comes into the room Teres doesn´t know that she has been screaming, but it often hurts when she turns over so that is probably why she screams. She is now being given laser treatment which seems to be quite successful in relieving the pain. Twice a month Teres swims in a swimming pool, once as part of her physiotherapy treatment, and once with her parents. This year she has not gone swimming because her arms hurt.

4.2.4 Teres' handicap aids

About three weeks ago, Teres' parents bought her a **water bed**, which has been a great success. It is comfortable to lie on and rest her back very well. The nursing table in her room is an ordinary desk, which her mother covers with a cushion.

Teres has a **Permobil** which her mother and father have painted in her favourite colour. The staff at the handicap aids centre did not like this at all, but her parents insisted. They had to sign a certificate promising to repaint the chair in its original colour if they returned it. Teres also has a manual wheelchair. Although it is called a junior wheelchair, it is a fairly large model. This wheelchair has a specially-made fibre cushion. The wheelchair is not height-adjustable but, in addition to the cushion, it has a specially-shortened seat and foot supports. The parents have made these

themselves because there are long delays for work to be done on modifications. The chair has also been repainted in lilac with pink arm rests.

The **Mulle wheelchair** is fitted with Teres´ own seat. This allows her to half-lie in the chair. In the winter the wheelchair is fitted with runners, which is an important modification for this family because they like to lead an outdoor life and even go fishing.

Teres has a **whirlpool bath.** It is in fact a massage cushion which produces air bubbles, and was bought with money from the grant.

Taking a shower is difficult for Teres. The family have been given a stretcher trollery to which they have attached a netting support so that it serves as a shower table. This apparatus is kept in the wash room.

Teres has a small **tricycle** for exercising. She only has the handlebars and the pedals, and she lies on her cushion and cucles to strengthen her feet and legs. This was a tip given her by the physiotherapist and she enjoys this training.

4.2.5 Handicap aids - unsolved problems

Teres has a typewriter at school, but this is not at all suitable because her hands are so week. She has supports on her arms and also wears a corset so it is difficult for her to reach all the keys.Teres has tried out a **Communicator** as a writing aid but her hands and arms are to weak to use it.

Teres cannot go to the lavatory herself. She urinates in an incontinence pad, but needs apparatus installed to help her position herself over the lavatory in a half-lying position. Teres needs something to enable her to sit on the floor. It would also be a good idea to have a two-way radio fitted to her Permobil. She is beginning to want to move outside the home far more now, and it is important for her not to have to worry about her wheelchair; it switches itself off all the time and Teres´ hands are not strong enough to enable her to switch it on again.

4.3 A meeting with Tommy

Tommy is five years old. He lives in a Stockholm suburb with his parents and a sister who is one year older. As soon as Tommy was born the doctor told his parents that he was suffering from osteogenesis imperfecta. They were given a fairly good picture of the implications of this disability. Because the condition was diagnosed immediately, the parents have never been under suspicion of child abuse. After about a year the family were put in touch with another family who had a child suffering from osteogenesis imperfecta.

Tommy is integrated into a day nursery but apart from the friends he has there he has no playmates who visit him at home. When he plays with other children his parents are often worried about fractures. Tommy is only worried himself if the activities become violent, and if they do, he calls for help. Most of Tommy's problems are related to clothing and the modifications of the environment in which he lives.

In Tommy´s case the technical aids he uses are standard apparatus which has been modified. The aids provided to the family have worked well. However, the family

does not know if they have been given all the aids they really need because they have not enough information of aids available. The most urgent problems they have in this area are related to leisure time activities. The family would very much like to be able to go for walks in the woods in the summer and be able to ski in the winter.

Most of the family´s needs in terms of handicap aids are of a general nature, and many of the appliances needed can be used by other children. Sadly, the professionals at the handicap aids centre do not understand the problems children suffering from osteogenesis imperfecta face. They have very little experience of this disability, says Tommy´s mother. The parents do not think that it is up to them to work out what aids their children need, and claim that the centre should be more helpful. "We don´t even know what is available!", they say. "Someone should be responsible for making sure that our needs in this area are met and then follow up the matter to make sure that, after modification and testing, the apparatus works well in the home environment.

Over the last year Tommy has broken five bones, and in the years before that he had a total of 12 fractures. To strenghten his muscles, Tommy trains in a public swimming pool. This is not a very satisfactory arrangement. The family would like to have a pool of their own at home, but this is impossible without a home modification grant.

4.3.1 Home modifications

Tommy's family lives in a detached house. When they were expecting Tommy they were offered a plot of land in the neighbourhood, which they took. The house is located high on the top of a hill and it is difficult to get up to it in a wheelchair. A few days after Tommy was born they were told that he had osteogenesis imperfecta, and although they say they were given a good description of what this disability involved, it wasn´t until 6 months later that they realized what the full implications were; for example, that Tommy would never be able to walk.

The family spoke to the local authority about getting another plot of land but the authorities were not in a position to give the family any priority in the queue for building land. So the only thing they could do was to modify their original house. They were not given any real assistance in modifying their home, and they had to do what they could themselves to plug the gaps as they occured. Today an urgent need is to modify the driveway up to the house. An extra entrance must also be built next to a veranda door and fitted with a lift because Tommy cannot use the main entrance. Part of the steep slope up to the house is to be surfaced with asphalt, and a veranda will be built at the back of the house to give a flat surface easily accessible for Tommy´s wheelchair. However, the family has now been given a new plot of land by the local authority, and are on the point of building a new home.

4.3.2 A day with Tommy

When Tommy wakes up in the morning his mother lifts him out of his bed and carries him over to the nursing table which is located above the bathtub. Tommy sleeps in a cot, but his parents want to replace it with a "proper" bed. They are very worried that he might fall out of his bed or try to get up himself and suffer bone fractures, and for this reason they have put off making the final decision on this matter.

When Tommy is dressed he sits in his Tripp-Trapp feeding chair and has breakfast. He has just been give a cushion to fit this chair, which is cut away on one side to

correct his incipient scoliosis. After the morning routines have been completed, Tommy´s parents drive him to the day nursery, where Tommy and his sister spend their days. Tommy is in a selection which takes children between 0 and 7 years old, but today the youngest child in this section is 3. There are 16 children in the group, that being the number of children the local authority has decided to have in all groups of children at day nurseries. A "resource" staff person has been employed for 40 hours a week to make things easier for Tommy. This job is divided between two people, one working 30 hours and the other 10 hours.

The day nursery opened two years ago. It is a new building designed to accomodate children with disabilities. Not withstanding this, parts of the bathroom had to be torn down and rebuilt, and this work took about a year and a half. There is no separate store for Tommy´s handicap aids, which are kept in Tommy´s nursing room. Britt, who looks after Tommy most of the time, says that the staff often complain that his apparatus is in the way. It does take up a lot of room, but is essential if Tommy´s life is to run smoothly. She also says that everyone working in the Tommy´s group knows how to lift him, and information has been circulated to all the departments in the day nursery about the proper way to life him. Many of the staff are afraid, and do not dare to lift him, but Britt says they have to learn because everyone must be able to help Tommy in an emergency. As a rule, people´s fear and uncertainty disappears once they have tried lifting him.

There are sometimes difficulties in the contacts with the other children. One day a little girl tried to lift Tommy - not to tease him or because she wanted to harm him, but because she wanted to mother him. Tommy screamed as loud as he could because that is his only chance to get away. He cannot put up any restistance and he is very frightened that someone will drop him on the floor, because he knows what will happen then. However, he has in fact only had one fracture at the day nursery, and that was when a child threw a soft cushin at him. Britt lets Tommy spend as much time as possible with the other children, and by now they now know what will injure Tommy and therefore avoid this.

4.3.3 Tommys interests

Like other preschool children Tommy is interested in playing with his friends, building things with lego and watching TV. Spending all day at the day care centre means that Tommy takes part in a wide variety of stimulating activities.

4.3.4 Tommy's handicap aids

Tommy has a **Brio Rehab wheelchair.** it is broken, and because the wheels are at an angle there is a risk that he will catch his fingers. It has been subjected to heavy wear and tear and, according to his parents, has never worked particulary well.

He also has a **Brio "Rolls Royce"** reclining wheelchair with a foot support. This works well, and it is fitted with a little **Nomeco** table which also works perfectly. The table is low, and Tommy can sit in the wheelchair and play at the table. This equipment serves its purpose well.

Tommy has a **Puky tricycle** with converted pedals fitted with foot straps to stop his feet slipping off the pedals. Unfortunately this is not very effective.

A Mothercare **baby bouncer** is hung in a doorway close to a large mirror on the wall which has been borrowed from Lekoteket, the toy library. Swinging in it is one of the things Tommy enjoys most.

Tommy has the following apparatus to help him at the day nursery.

A **Comfort Micro** electric wheelchair. It is light blue, and he likes it very much. He finds it difficult to back the wheelchair without running into something. He has cushions in the back support which makes it difficult to watch out behind, and this increases the risk that he will run into other children. Fitting the wheelchair with rear view mirrors has been discussed, but the physiotherapist is not in favour of this because Tommy will then no longer practice turning around.

Tommy also has a manual wheelchair at the day nursery. It is a little **Swede 20** and it goes very fast. Tommy also has a **wheeled walking frame**, a **Bobbath ball**, and an **Airex gymnastic mat** at the day nursery. He also has the same type of Tripp-Trapp chair that he has at home, with a specially designed cushion.

4.3.5 Handicap aids - unsolved problems

The position of the house and modifications to the house are two of the most urgent problems for the family .However, these difficulties will disappear when the family builds the new house on the plot provided by the local authority. There are many difficulties related to the modification of the home. Tommy is still young and new problems will have to be faced and attended to as they arise.

5. CONCLUSION

This survey shows that families who have children with osteogenesis imperfecta need more support from the community than they are given today.

The diagnosis of this condition is an important issue for the parents and it has to be made far earlier than is sometimes the case today. This is particulary important in order to protect the parents from suspicions of child abuse. To note is that this has been the case for 11 of the 24 families covered in this survey. Often the parents suspect that there is something wrong with their children early in life, which gives them a lot of worry. However, with the additional suspicion of child abuse they are subjected to a considerable increase in emotional pressure. Because the small number of children born each year with osteogenesis imperfecta profesionals have relatively little experience of this condition. More information and training is required on this disability, its symptoms, and future implications.

When it comes to handicap aids, the survey indicates that there are many needs which have not yet been met. The major problem with handicap aids is that it is difficult to find any equipment which serves its purpose well in everyday life. Not only have the parents complained about the supply of handicap aids, they have also provided detailed descriptions of their need for apparatus which will require both new

development and special modifications. The parents feel that they are given inadequate information about the equipment currently available. The greatest worry for the parents is the glaring ignorance about children with osteogenesis imperfecta., A constantly recurring question is how information can be disseminated spread to professionals working with handicap aids and in the care sector.

Thus there are major shortcomings in the field of information and education on handicap aids and on the modification of those aids, and more resources are needed in this area. Several parents said that they had no way of knowing whether they had been given the apparatus they needed because they did not know what was available. The purpose of these aids is to compensate for, and provide support for, a reduced capacity, so that a disability is avoided or the effects of a disability are reduced. Functional aids will enable these children to become more independent and thus enjoy a richer life. There is a need today to develop equipment which makes daily nursing care easier and to make it possible for these children to enjoy the companionship of other children. Moreover, it is important that an evaluation is made of the equipment already prescribed for use in the home.

A lack of understanding of the implications of this disability has also meant that the families have not been given proper assistance in modifying their homes. The families felt that they had been deserted by the authorities and that they had to find ways of solving their problems themselves. At times this meant that they resolved the problem in a way that was unsatisfactory in the long run, and the work they had done had to be done again. It is therefore essential that knowledge gained from successful home modifications is documented and spread.

Hydrotherapy is very beneficial to the muscles, and most of the children had a very positive attitude towards this activity. Many of the families had access to a pool, but only three families had a pool in the home. In the summer of 1986 the Supreme Administrative Court dealt with the issue of whether installing pools in houses could be classified as a home modification for the disabled. The Court ruled that this was the case, a judgement which will serve as a precedent in other cases of this nature.

It is also worth noting that only 14 of the 24 children have playmates. It is highly probable that those children often are left to play on their own due to their anxiety about the risk of fractures. In addition, half of the parents admit that they are often worried when their children play with friends, and the rest of the parents say they are worried sometimes. Only four parents said that they thought their children were worried about breaking bones when they were at play. This shows that the parents are more worried than their children about fractures. There is a risk that parents who live with the daily fear that their children will be injured can inhibit the development of the child. This is a difficult issue for the parents, and hardly one which can be resolved by means of counselling from "experts" on the development of children. There is no simple advice to give which would help these parents. They must come to the realization in their own pace that they can diminish their attachment and hold on their children. In this the parents will be guided by the degree of maturity of their children and their awarness of their disability. Maria is a good example of this. She is a typical teenage girl who knows what she wants and dares to speak her mind to her parents. She is open, secure and extravert, and she knows both what she is capable of doing and her limitations. Even Teres, who is only eight, is showing signs of being aware of her disability. Tommy, too, knows the limits imposed by his condition. To give an example, when another child lifts him up, he screams for a member of the staff to come and help. He knows what will happen if he falls on the floor.

Under the Social Services Act, parents of disabled children are entitled to help in the home to give them some time for themselves, for their other children or to run errands and meet friends. It is very difficult for families with children suffering from osteogenesis imperfecta to make use of this service, because other people have so little knowledge and experience of this particular disability. One of the first steps on the way to give these families the help they need to live under the same conditions as others is to increase the amount of information and knowledge available about the children´s disability. This will increase people´s understanding of the children´s and their families´ circumstances.

On a number of occasions the associations for osteogenesis imperfecta sufferers in the Nordic countries have discussed the idea of a centre where the families and their children can go for diagnosis, treatment, surgery and to test handicap aids, that is to say, a highly specialized centre which can operate on a Nordic basis. Because this is such a small group of disabled people there is a need for a centre which offers qualified help from experts in all the sub-areas involved.

If children with osteogenesis imperfecta are to enjoy optimal development and live as normal a life as possible, then they and their families must be given sufficient support by the community.

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